Journal of Chemical Health Risks

www.jchr.org

JCHR (2024) 14(4), 1119-1123 | ISSN:2251-6727

Wilkie's Syndrome: A Bizarre Adventure into Abdomen

[1]Dr. Shreethi.S. Pillai, [2]Prof Dr.Balaji.D

[1]Post Graduate, [2]Professor, SRM Medical College Hospital And Research Centre,Potheri,Kattankulathur

(Received: 14 April 2024 Revised: 1 May 2024 Accepted: 18 June 2024)

KEYWORDS

Wilkie's syndrome, SMA, postprandial.

ABSTRACT:

Introduction: Wilkie's Syndrome (also called SMA Syndrome) is a rare cause of small bowel obstruction, characterized by an extrinsic vascular compression of the third portion of the duodenum between the abdominal aorta and overlying superior mesenteric artery, due to loss of cushion of fat.

Case Presentation : We describe the case of a 62yr old female patient who presented with upper abdominal distension and repeated episodes of vomiting who was diagnosed as a case of Wilkie's syndrome on radiological investigation. Emergency laparotomy with duodenojejunostomy was done.

Conclusion: Wilkie's Syndrome is a rare disorder. Only 500 cases reported worldwide till date. Surgical treatment is the treatment of choice in long standing cases otherwise it may result in fatal complications like advanced malnutrition, electrolyte imbalance, gastric rupture, spontaneous upper GI bleeding or sudden cardiovascular collapse.

1. Introduction

Superior mesenteric artery (SMA) syndrome, also called Wilkie's syndrome or cast syndrome, is a rare disorder in which acute angulation of the SMA causes compression of the third part of the duodenum between the SMA and the aorta, leading to obstruction. SMA syndrome is an atypical cause of proximal intestinal obstruction, most frequently occurring in young patients who have had an important weight loss. Surgeries for spinal deformities as well as high insertion of the ligament of Treitz are other potential causes for the occurrence of SMA syndrome [1,2]. Loss of retroperitoneal fatty tissue as a result of this variety of conditions is believed to be the etiologic factor causing the acute angulation. Symptoms vary from postprandial nausea and bilious vomiting to abdominal pain as well as weight loss and can occur acutely or chronically [3]. The severity of the symptoms largely depends on the degree of the compression as reflected by the aortomesenteric angle.

SMA syndrome was first described by the Austrian professor Carl Freiherr von Rokitansky in 1861 as an autopsy finding [4]. Later, Wilkie provided a more detailed clinical and pathophysiologic description in a series of 64 patients and suggested treatment approaches [5]. After that, a controversy regarding the actual existence of this syndrome started, especially because of the lack of specificity of the symptoms and the long list of differential diagnoses. However, advances in imaging, such as in computed tomography (CT) and magnetic resonance imaging, have tremendously helped with clear visualization of the angle between the aorta and the SMA and thus improved the diagnostic rate [6]. In adults, clinical SMA syndrome manifestations appear if the angle drops below 20°, and it is believed that values of this angle may be lower for paediatric patients [5]. Thus, in the appropriate clinical context, detailed history as well as imaging findings should highly raise the clinical suspicion for the diagnosis of SMA syndrome. A delay in this diagnosis can potentially lead to many complications, such as electrolyte imbalance, catabolic wasting, peritonitis and gastric perforation.

Conservative therapy mainly consists of weight gain achieved orally or parenterally, with the aim of restituting the mesenteric fat pad and increasing the aortomesenteric angle [7]. If this non-invasive approach fails, surgical therapy may be the next approach, with duodenojejunostomy being the currently preferred treatment [8].

Case Presentation: A 62year old female patient presented to the emergency department with upper abdominal distension and repeated episodes of vomiting for 1 day. Vomiting occurs after 3-4 hours of taking

Journal of Chemical Health Risks
www.jchr.org
JCHR (2024) 14(4), 1119-1123 | ISSN:2251-6727

mealwhich was bilious, non-projectile containing eaten food particles. Vomiting occurs most of the time after taking solid meal. There was no history of constipation. History of weight loss was present in four months (70kg to 55kg). No history of any trauma.

Past history: Not a known case of type 2 diabetes mellitus, systemic hypertension, thyroid disorders, seizures

Personal History: Patient followed a mixed diet with normal bowel and bladder habits
General Physical Examination: Patient was conscious and oriented to time, place and person. Blood pressure - 110/70mmHg.
Pulse rate – 86/min.
Respiratory rate – 16/min.

Temperature – Afebrile.
Built – Lean thin.
No Icterus, No Cynosis, No Clubbing and No any lymphadenopathy. Pallor – Present.

Systemic Examination: Abdomen – On inspection - distended.
On palpation – Soft, distended, no guarding, no rigidity, no tenderness, no palpable lump.

On percussion – Tympanic note.
On Auscultation – No bowel sounds present.

Radiological Investigations:

X-ray erect abdomen – Distended stomach with air fluid levels in the stomach and duodenum.

CECT abdomen (with IV contrast)-Superior mesenteric vessels compressing the 3rd part of duodenum with proximal dilatation – SMA Syndrome

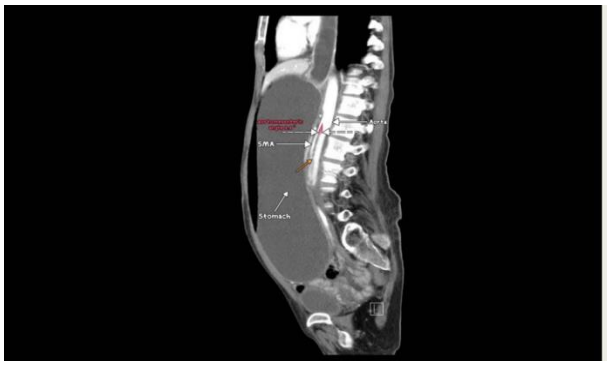

Figure 1: CECT Image Showing Compression Of 3rd Part Of Duodenum Between SMA And Aorta

Operative Management: Exploratory laparotomy was done. Grossly dilated stomach and proximal duodenum were seen. D3 segment was compressed between the underlying abdominal aorta and overlying SMA. Kocherization of duodenum was performed and jejunal loops 18-20cms from the duodeno jejunal flexure was selected for anastomosis. Hand sewn 2 layered duodeno-jejunostomy was performed anterior to Superior mesenteric artery (no tension or anastomotic leak were noticed).

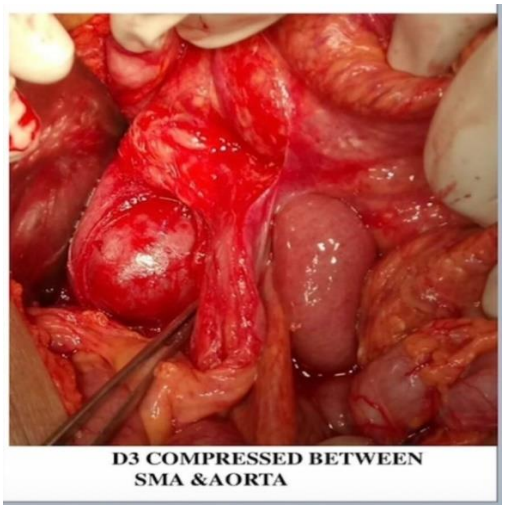

Figure 2: Intraoperative Picture Showing D3 Compressed Between Sma And Aorta

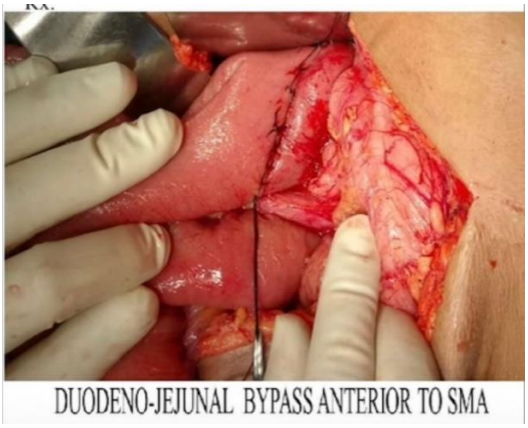

Figure 3: Intraoperative Picture Showing Duodenojejunostomy

Discussion

This syndrome was described by Rokitansky more than 100 years ago in 1861.[8]It is extremely rare, and an incidence of 0.2% was found in 6000 radiological

Journal of Chemical Health Risks

www.jchr.org

JCHR (2024) 14(4), 1119-1123 | ISSN:2251-6727

studies. Three duo- denal obstruction mechanisms have been described: (1) a very acute aortomesenteric angle; (2) a transversal duode- num in a high position due to a short Treitz ligament, and (3) an anomalous route of the mesenteric artery or one of its branches, displaced downwards and in front of the spinal column.[14] This condition is more common in women and young adults, and in the majority of cases it presents after a major weight loss. Nevertheless, in 2006 Biank and Werlin[21] published a series of 22 children, of whom only 50% had lost weight prior to their diagnosis. The causes of Wilkie syndrome may be classified in five groups: consumptive syn- dromes (AIDS, cancer, major burns, endocrinal pathologies, poor intestinal absorption); eating disorders (anorexia ner- vosa); postoperative (orthopaedic surgery, spinal surgery); severe trauma (craneoencephalic trauma, polytraumatism) and spinal deformities, diseases or lesions. Cases have been published recently in association with rapid weight loss after bariatric surgery.[15]

Wilkie syndrome symptoms are non-specific, and it may present as intolerance to food with nausea and vomiting, weight loss, early satiety, abdominal distension and epi- gastric pain. The pain may be relieved when the patient lies in prone decubitus, left lateral decubitus or a genupec- toral position. These manoeuvres relax the pressure of the mesenteric artery on the duodenum,[13] and this may guide diagnosis.

Patients may complain about reflux, which may be shown by the endoscopic study of stasis-linked esophagitis or gas- tritis. There is also a higher prevalence of duodenal ulcers than is the case for the general population (up to 45%). Fatality is secondary to severe hydroelectric alterations, gastric perforation, an obstructive bezoar and gastric or por- tal pneumatosis. Once the clinical symptoms are established they self-perpetuate, regardless of their aetiology.[12]

Historically a barium study that made it possible to observe the dilation of the first and second duodenal por- tions and the compression of the third portion was the diagnostic test used, together with arteriography. Never- theless, computerised angiotomography has recently been shown to have greater diagnostic sensitivity.

The severity of the symptoms is correlated with aor- tomesenteric distance. Endoscopy helps to rule out intrinsic lesions of the digestive tract that may lead to obstruction, and it also permits taking biopsies.[16]

In acute or mild cases, conservative treatment should be attempted first. Nasogastric tube placement for duodenal and gastric decompression and mobilization into the prone or left lateral decubitus position often is effective in the acute setting.[19]

Acute superior mesenteric artery syndrome involving the reversal or removal of the precipitating factor with proper nutrition and replacement of fluid and electrolytes, either by surgically inserted jejunal feeding tube, nasogastric intubation or peripherally inserted central catheter (PICC line) administering total parenteral nutrition (TPN). Pro- motility agents such as metoclopramide may also be beneficial.[18]

If conservative treatment fails, or if the case is severe or chronic, surgical intervention is required. The most common operation for SMA syndrome, duodenojejunostomy. Bypassing the compression caused by the AA and the SMA.[1]

Less common surgical treatments for SMA syndrome include Roux-en-Y duodenojejunostomy, gastro- jejunostomy, and anterior transposition of the third portion of the duodenum, intestinal de rotation, and division of the ligament to Treitz (Strong's operation).

The possible persistence of symptoms after surgical bypass can be traced to the remaining prominence of reversed peristalsis in contrast to direct peristalsis, although the precipitating factor (the duodenal compression) has been bypassed or relieved. Reversed peristalsis has been shown to respond to duodenal circular drainage—a complex and invasive open surgical procedure originally implemented and performed in China.

This syndrome was described by Rokitansky more than 100 years ago in 1861.[8]It is extremely rare, and an incidence of 0.2% was found in 6000 radiological studies. Three duo- denal obstruction mechanisms have been described: (1) a very acute aortomesenteric angle; (2) a transversal duode- num in a high position due to a short Treitz ligament, and (3) an anomalous route of the mesenteric artery or one of its branches, displaced downwards and in front of the spinal column.[14] This

Journal of Chemical Health Risks

www.jchr.org

JCHR (2024) 14(4), 1119-1123 | ISSN:2251-6727

condition is more common in women and young adults, and in the majority of cases it presents after a major weight loss. Nevertheless, in 2006 Biank and Werlin[21] published a series of 22 children, of whom only 50% had lost weight prior to their diagnosis. The causes of Wilkie syndrome may be classified in five groups: consumptive syn- dromes (AIDS, cancer, major burns, endocrinal pathologies, poor intestinal absorption); eating disorders (anorexia ner- vosa); postoperative (orthopaedic surgery, spinal surgery); severe trauma (craneoencephalic trauma, polytraumatism) and spinal deformities, diseases or lesions. Cases have been published recently in association with rapid weight loss after bariatric surgery.[15]

Wilkie syndrome symptoms are non-specific, and it may present as intolerance to food with nausea and vomiting, weight loss, early satiety, abdominal distension and epi- gastric pain. The pain may be relieved when the patient lies in prone decubitus, left lateral decubitus or a genupec- toral position. These manoeuvres relax the pressure of the mesenteric artery on the duodenum,[13] and this may guide diagnosis.

Patients may complain about reflux, which may be shown by the endoscopic study of stasis-linked esophagitis or gas- tritis. There is also a higher prevalence of duodenal ulcers than is the case for the general population (up to 45%). Fatality is secondary to severe hydroelectric alterations, gastric perforation, an obstructive bezoar and gastric or por- tal pneumatosis. Once the clinical symptoms are established they self-perpetuate, regardless of their aetiology.[12]

Historically a barium study that made it possible to observe the dilation of the first and second duodenal por- tions and the compression of the third portion was the diagnostic test used, together with arteriography. Never- theless, computerised angiotomography has recently been shown to have greater diagnostic sensitivity.

The severity of the symptoms is correlated with aor- tomesenteric distance. Endoscopy helps to rule out intrinsic lesions of the digestive tract that may lead to obstruction, and it also permits taking biopsies.[16]

In acute or mild cases, conservative treatment should be attempted first. Nasogastric tube placement for duodenal and gastric decompression and mobilization into the prone or left lateral decubitus position often is effective in the acute setting.[19]

Acute superior mesenteric artery syndrome involving the reversal or removal of the precipitating factor with proper nutrition and replacement of fluid and electrolytes, either by surgically inserted jejunal feeding tube, nasogastric intubation or peripherally inserted central catheter (PICC line) administering total parenteral nutrition (TPN). Pro- motility agents such as metoclopramide may also be beneficial.[18]

If conservative treatment fails, or if the case is severe or chronic, surgical intervention is required. The most common operation for SMA syndrome, duodenojejunostomy. Bypassing the compression caused by the AA and the SMA.[1]

Less common surgical treatments for SMA syndrome include Roux-en-Y duodenojejunostomy, gastro- jejunostomy, and anterior transposition of the third portion of the duodenum, intestinal de rotation, and division of the ligament to Treitz (Strong's operation).

The possible persistence of symptoms after surgical bypass can be traced to the remaining prominence of reversed peristalsis in contrast to direct peristalsis, although the precipitating factor (the duodenal compression) has been bypassed or relieved. Reversed peristalsis has been shown to respond to duodenal circular drainage—a complex and invasive open surgical procedure originally implemented and performed in China.

Conclusion: SMA is a rare disorder which is mostly common among adolescents and young adult females. It can present as chronic, intermittent or acute intestinal obstruction. A delay in diagnosis can lead to complications such as electrolyte imbalance, catabolic wasting, peritonitis and gastric perforation. Initial treatment is always conservative- adequate nutrition, hydration with intravenous fluids and nasogastric decompression. Laparoscopic/open duodeno jejunostomy is the surgery of choice in acute obstruction or failed medical treatment.

References:

1. Strong EK: Mechanics of arteriomesenteric duodenal obstruction and direct surgical attack upon etiology. Ann Surg 1958;148:725-730.

Journal of Chemical Health Risks

www.jchr.org

JCHR (2024) 14(4), 1119-1123 | ISSN:2251-6727

2. Sapkas G, O'Brien JP: Vascular compression of the duodenum (cast syndrome) associated with the treatment of spinal deformities. A report of six cases. Arch Orthop Trauma Surg 1981;98:7-11.
3. Record JL, Morris BG, Adolph VR: Resolution of refractory superior mesenteric artery syndrome with laparoscopic duodenojejunostomy: pediatric case series with spectrum of clinical imaging. Ochsner J 2015;15:74-78.
4. von Rokitansky C: Lehrbuch der pathologischen Anatomie, vol 3, ed 3. Vienna, Braumüller und Seidel, 1861, p 87.
5. Wilkie D: Chronic duodenal ileus. Am J Med Sci 1927;173:643-649.
6. Unal B, Aktaş A, Kemal G, Bilgili Y, Güliter S, Daphan C, Aydinuraz K: Superior mesenteric artery syndrome: CT and ultrasonography findings. Diagn Interv Radiol 2005;11:90-95.
7. Mandarry M, Zhao L, Zhang C, Wei Z: A comprehensive review of superior mesenteric artery syndrome. Eur Surg 2010;42:229-236.
8. Rokitansky CV. Lehrbuch der patologischen Anatomie. 3rd ed. Vienna: Braumuller; 1861. p. 187.
9. Zhu ZZ, Qiu Y. Superior mesenteric artery syndrome following scoliosis surgery: its risk indicators and treatment strategy. World J Gastroenterol. 2005;11:3307-10.
10. Thomas JS, Wayne SC, Michael DL, Shanu NK, La CW. Supe- rior mesenteric artery syndrome after laparoscopic Roux-en-Y gastric bypass. Surgery. 2005;137:383-5.
11. Benjamin C, Bruce A. Superior mesenteric artery syndrome after Roux-en-Y gastric bypass. JSLS. 2010;14:143-6.
12. Richardson WS, Surowiec WJ. Laparoscopic repair of superior mesenteric artery syndrome. Am J Surg. 2001;181:377-8.
11. Merrett ND, Wilson RB, Cosman P, Biankin AV. Superior mesen- teric artery syndrome: diagnosis and treatment strategies. J Gastrointest Surg. 2009;13:287-92.
12. Akin JT Jr, Skandalakis JE, Gray SW. The anatomic basis of vascular compression of the duodenum. Surg Clin N Am. 1974:1361-70.
13. Hines RH, Gore RM, Ballantyne GH. Superior mesenteric artery syndrome: diagnostic criteria and therapeutic approaches. Am J Surg. 1984;148:630-2.
14. Mansberger AR Jr, Hearn JB, Byers RM, Feisig N, Buxton RW. Vascular compression of the duodenum. Emphasis on accurate diagnosis. Am J Surg. 1968;115:89-96.
15. Makam R, Chamany T, Potluri VS, Varadaraju PJ, Murthy R. Laparoscopic management of superior mesenteric arter
16. Wilkie DP. Chronic duodenal ileus. Br J Surg. 1921;9:204.
19. Derrick JR, Fadhli HA. Surgical anatomy of the superior mesenteric artery. Am Surg. 1965;31:545-7.
20. Fernández López MT, Lop´ez Otero MJ, Bardasco Alonso ML, Álvarez Vázquez P, Rivero Luis MT, López Barros G. Sín´drome de Wilkie: a propósito de un caso. Nutr Hosp. 2011;26:646-9. 9. Cohen LB, Field SP, Sachar DB. The superior mesenteric artery syndrome. The disease that isn't, or is it? J Clin Gastroenterol. 1985;7:113-6.
21. Biank V, Werlin S. Superior mesenteric artery syndrome in children: a 20-year experience. J Pediatr Gastroenterol Nutr. 2006;42:522-5.